Idiopathic Pulmonary Fibrosis

Exh D

What is Idiopathic Pulmonary Fibrosis?

Idiopathic Pulmonary Fibrosis (IPF) is a disease of inflammation that results in scarring, or fibrosis, of the lungs. In time, this fibrosis can build up to the point where the lungs are unable to provide oxygen to the tissues of the body.

Doctors use the word "idiopathic" (from the Greek "idio" meaning "peculiar" or "unusual" and "pathy" meaning "illness") to describe the disease, because the cause of IPF is unknown. Currently, researchers believe that IPF may result from either an autoimmune disorder, a condition in which the body's immune system attacks its own tissues, or the after effects of an infection, most likely a virus.

Whatever the trigger is for IPF, it appears to set off a series of events in which the inflammation and immune activity in the lungs -- and, eventually, the fibrosis processes, too--become uncontrollable. In a few cases, heredity appears to play a part, possibly making some individuals more likely than others to get IPF.

In studies of patients with IPF, the average survival rate has been found to be 4 to 6 years after diagnosis. Those who develop idiopathic pulmonary fibrosis at a young age seem to have a longer survival.

Who Gets IPF

The exact number of people who develop idiopathic pulmonary fibrosis each year is not known. It is known, however, that equal numbers of men and women get the illness and that most cases of IPF are diagnosed when the patients are between the ages of 40 and 70.

Symptoms

Early symptoms of idiopathic pulmonary fibrosis are usually similar to those of other lung diseases. Very often, for example, patients suffer from a dry cough and dyspnea (shortness of breath). As the disease progresses, dyspnea becomes the major problem. Day-to-day activities such as climbing stairs, walking short distances, dressing, and even talking on the phone and eating become more difficult and sometimes nearly impossible. Enlargement (clubbing) of the fingertips may develop. The patient may also become less able to fight infection. In advanced stages of the illness, the patient may need oxygen all the time.

IPF can lead to death. Often the immediate cause is respiratory failure due to hypoxemia, right-heart failure, a heart attack, blood clot (embolism) in the lungs, stroke, or lung infection brought on by the disease.

The Course Of IPF

Although the course of idiopathic pulmonary fibrosis varies greatly from person to person, the disease usually develops slowly, sometimes over years.

The early stages are marked by alveolitis, an inflammation of the air sacs called alveoli, in the lungs. The job of the air sacs is to allow the transfer of oxygen from the lungs into the blood and the elimination of carbon dioxide from the lungs and out of the body.

As IPF progresses, the alveoli become damaged and scarred, thus stiffening the lungs. The stiffening makes breathing difficult and brings on a feeling of breathlessness (dyspnea), especially during activities that require extra effort.

In addition, scarring of the alveoli reduces the ability of the lungs to transfer oxygen. The resulting lack of oxygen in the blood (hypoxemia) may cause increases in the pressure inside the blood vessels of the lungs, a situation known as pulmonary hypertension. The high blood pressure in the lungs then puts a strain on the right ventricle, the lower right side of the heart, which pumps the oxygen-poor blood into the lungs.

How IPF is Diagnosed

The first suspicion that a person may have idiopathic pulmonary fibrosis is usually based on the patient's symptoms and medical history. The doctor will try to confirm or rule out any suspicion by ordering one or more of the following tests:

Chest x-ray
A simple chest x-ray is a picture of the lungs and surrounding tissues, most often taken while the patient is standing up. In an IPF patient, the x-ray usually reveals shadows, mostly in the lower part of the lungs. In addition, lung size tends to appear smaller than normal.

Computed Tomography (CT)
A computed tomography scan of the chest is a series of x-rays that provide a view of the lungs that looks almost as if a slice had been made through the chest. During a CT scan, the patient lies inside a long, oval-shaped machine that permits x-ray beams to pass through the top, sides, and back of the body. A computer is used to combine all the pictures taken from these positions and thus gives the doctor a good look at what's going on inside the lungs and chest.

Blood Tests
When IPF is suspected, the doctor will analyze the patient's blood. A low level of oxygen in the arterial blood may reveal that the alveoli are not taking up enough oxygen.

Pulmonary Function Tests
Pulmonary function tests (PFTs) require the patient to breathe into a mouthpiece. The mouthpiece, in turn, is connected to a machine that measures the amount of air the patient breathes in and out over a specific period of

time. The results tell the doctor how well the air passages in the lungs are functioning and how well the lungs are expanding.

Bronchoalveolar Lavage

Lung washings (bronchoalveolar lavage) are also helpful in arriving at a diagnosis of IPF. In this procedure, the doctor inserts a long, narrow, flexible, lighted tube called a bronchoscope down the windpipe and into the lungs to remove fluid (lavage) and other materials from inside the lungs.

Even if some or all of the results from such tests are abnormal, they are rarely sufficient to make a specific diagnosis of IPF. The only way the doctor can confirm a diagnosis of IPF is by examining the lung tissue; such tissue is usually obtained by an open lung biopsy.

Open Lung Biopsy

In an open lung biopsy, a chest surgeon makes cuts between the ribs in the chest and removes small pieces of tissue from several places in the lungs. The material is examined in the laboratory to determine how much inflammation and fibrosis are in the lungs. It is the only way to confirm whether the patient has IPF. If IPF is present, the biopsy results are also the best way to find out how far
the disease has progressed and what the outlook is.

In a patient with no other significant illness, recovery from an open lung biopsy is relatively quick. The hospital stay is usually 4 to 7 days; some newer procedures require less surgery, bringing hospital stays to 1 to 3 days.

Treatment

The best chance of slowing the progress of IPF is by treatment as soon as possible. Most IPF patients require treatment throughout life, usually under the guidance of a lung specialist. Some major medical centers and large teaching hospitals do research on the disease and provide consultation and treatment to patients.

Treatment for idiopathic pulmonary fibrosis may vary a great deal. It depends on many things, including the age of the patient and stage of the disease. The aim of treatment is to reduce the inflammation of the alveoli and stop the abnormal process that ends in fibrosis. Once scar tissue has formed in the lung, it cannot be returned to normal.

Drugs are the primary way that IPF is treated. They are usually prescribed for at least 3 to 6 months. This gives the doctor time to see if a particular treatment is effective. A combination of tests is used to monitor how well a particular drug is working. The dose may have to be adjusted so that the medicine gives the best

possible results with the least side effects. Most side effects are reduced when the dose is made smaller or the drug is stopped. Commonly used drugs are prednisone and cytoxan. Oxygen administration and, in special cases, transplantation of the lung are other choices.

Prednisone

A corticosteroid, prednisone, is the most common drug given to patients with idiopathic pulmonary fibrosis. About 25 to 35 percent of all patients respond favorably to this medicine. No one knows exactly how corticosteroids work or why some patients do well on prednisone while others do not. Patients take prednisone by mouth every morning, starting with a high dose for the first 4 to 8 weeks. As they improve, they gradually take smaller amounts. Changes in mood are one of the more common side effects of prednisone; most patients, however, can handle the mood changes -- anxiety, depression, or sleeplessness -- once they know what is causing the problem. A less common side effect is a rise in blood-sugar levels, osteoporosis, high blood pressure, cataracts, and increased susceptibility to infection.

Cytoxan

Cyclophosphamide, also referred to as cytoxan, may be taken together with prednisone, or instead of it. Like prednisone, cytoxan is swallowed each day.

One of the more serious side effects of cyclophosphamide is leukopoenia, a condition in which the number of white blood cells drops to a dangerously low level. Leukopoenia can be controlled by regularly checking the blood count and adjusting the dose of cytoxan if necessary.

Other Medicines

Azathioprine, penicillamine, chlorambucil, vincristine sulfate, and colchicine have been used in a few patients with idiopathic pulmonary fibrosis. The newer treatments use drugs which are still in phase two trials such as: interferon-gamma 1b, and an antifibrotic agent, pirfenidone.

Oxygen

In addition to treatment with medicine, some patients may need oxygen, especially when blood oxygen becomes low. This treatment helps re-supply the blood with oxygen. As a result, breathlessness is reduced, the patient can be more active, and the severity of pulmonary hypertension decreases.

Exercise

Regular exercise may be useful for patients with IPF. A daily walk or regular use of a stationary bicycle or

treadmill can improve muscle strength and breathing ability and also increase overall strength. If needed, supplemental oxygen should be used; sometimes it is the only way a patient is able to do a reasonable amount of activity.

Lung Transplantation

Lung transplantation, either of both lungs or only one, is an alternative to drug treatment for patients in the severe, final stages of IPF. It is most often performed in patients under 60 years of age who do not respond to any form of treatment. The survival rate is approximately 60 percent.

Lifestyle

Many IPF patients, particularly those in the early stages of the disease, respond to drug treatment and can continue to go about most of their normal activities, including working. Some patients with advanced IPF need to carry oxygen with them.

In addition to getting proper treatment, IPF patients can help themselves by following the same sensible health measures that everyone should observe. These include eating a healthy diet, maintaining proper weight, exercising regularly, and getting enough rest. Above all, IPF patients should not smoke. Pregnancy is not advisable because the illness puts an extra load on the heart and lungs.

As with many chronic illnesses, emotional support and psychological counseling can be of much help to the patient. Most doctors and patients agree that it is important for both patient and family to be as informed as possible about IPF. In this way, everyone involved can understand the illness and apply that information to what is happening in his or her own life.

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